

FRIEDREICH'S ATAXIA; ITS RELATION TO CONDUCTING PATHS IN THE CORD.¹

BY DAVID INGLIS, M.D., OF DETROIT.

WITHIN the past year I had the opportunity of presenting a case of Friedreich's ataxia, of which I made a full report at the last meeting of the Michigan State Medical Society.

In brief the case is this :

When the boy was four years old he had an attack of measles from which he did not promptly recover. He was unable to walk for seven weeks, after which time he began to get about, but showed an unsteady gait and was easily pushed over. During two years, from this time on, the history was that he had had occasional pains, which were not severe, but resembled "growing pains." The boy's gait, as stated, became unsteady from the time that he got up from the measles. Before that time he had learned to walk at the usual age, and had seemed in every way a normal child. His gait grew steadily worse, so that when I saw him, two years after the onset of the trouble, he presented in perfection, the whole picture of ataxia. When he attempts to walk he keeps his feet widely separated, his head and body bent forward, his vision fixed on the floor and his arms spread and instinctively trying to touch every object as he passes in order to give some little aid in keeping his balance as he totters zigzag forward. He seems to be perpetually falling or as if about to run. When he attempts to stand or even to sit erect without some support to his back, he sways continually. So marked is the loss of steady motor control, that when sitting in a chair he constantly holds on to the seat with both hands. If he closes his eyes he sways still more and, when standing, falls. In short, he presents both static and locomotor ataxy. In using his arms and hands the same unsteadiness is manifest. He can, indeed, perform almost any act, but when, for example, he buttons his coat the action is clumsy and slow, and not only the hand but the forearm and shoulder muscles are all brought

¹ Read at the annual meeting of the American Neurological Society, Washington, D. C., September 22, 23 and 24, 1891.

into the effort. This inco-ordination of the upper extremities came on several months after the affection of the legs, and now, within the last two months, the inco-ordination has involved centres still higher up; for his speech has now become slow, monotonous and difficult. The mother claims that the boy's mind is as clear and bright as ever, but that he has difficulty in speaking.

We have, then, to deal with a defect of co-ordination of wide extent, one which involves the entire muscular control from the legs up as far as the tongue and pharynx.

Meanwhile the lad's general health is good; he is well nourished, has no muscular wasting; muscular strength is good.

The patellar reflex is wholly lost. Cremaster and pupillary reflexes normal. No defect of vision, no nystagmus.

As stated, the boy has had occasional trouble with aching pains, but examination of his sensory apparatus shows that it is in excellent condition. I have repeatedly tested him and find that his perception of touch, pain and temperature is both accurate and prompt.

Special senses normal; electrical reactions normal; no history of any impairment of his organic functions.

Such is the group of symptoms, and from it I have felt justified in classing the case as one of Friedreich's ataxia. I do so in spite of the fact that the hereditary character of the disease is not demonstrable. The boy is the oldest child, and no other case has occurred in the family. I should also state that his father is a man of good habits, and no neurotic inheritance can be elicited.

Friedreich in his original article summarized the clinical characters of the disease as follows:

"Impairment in the co-ordination and harmony of movements developing gradually and spreading from the lower to the upper half of the body, and always involving finally the organs of speech.

"Sensibility and the functions of the special senses and of the brain being intact, paralysis of the sphincters and trophic disturbances are absent; less constant phenomena

are, curvature of the spine, sensations of vertigo and nystagmus.

"From a clinical point of view we must regard the disease as a progressive paralysis of the faculty of combination of movements."

My case conforms accurately to Friedreich's description, and the absence of any family tendency up to the present time does not impair the classification.

My object, however, in reporting the case is not to discuss the differential diagnosis of Friedreich's disease, but to consider the bearing of the affection upon our views of the physiological anatomy of the function of co-ordination in the spinal cord.

There have been, so far, thirteen autopsies upon cases of Friedreich's ataxia: five by Friedreich, two by Rutimeyer and one each by Everett Smith, Pitt, Blocq, Letulle, Auscher and Menzel.

Pitt's description of the pathological changes in his case serves as a good basis of comparison, it is:

1st. The spinal cord is extremely slender.

2d. Extreme sclerosis of the columns of Goll in their whole length, from the lumbar enlargement to their termination in the floor of the fourth ventricle.

3d. Intense sclerosis of the posterior part of the columns of Burdach, in which, however, some healthy fibres are scattered.

4th. The zone of the columns of Burdach which bounds the corner and the posterior root is intact.²

5th. Sclerosis not distinctly limited (much less intense than in the posterior columns of the crossed pyramidal tracts.)

6th. Sclerosis of the ascending cerebellar tracts, visible up to the decussation of the pyramids. Very slight and irregular sclerosis of a few scattered fibres in the antero-lateral columns.

7th. Degeneration of the columns of Clarke in some sections.

² Marginal zone of Lissauer.

8th. Degeneration of some fibres of the posterior roots and of the posterior corona.

Ladame, in his summary of nine autopsies, shows that there is a general agreement with such a pathological picture as that of Pitt, although some autopsies show the sclerosis to be less widely spread. In Friedreich's first case only the posterior columns were affected. The four autopsies made during the past year also conform very closely to the description. The columns of Goll are uniformly most affected, then the posterior part of Burdach's, the column of Clarke and direct cerebellar also pretty uniformly, the crossed pyramidal to a much less degree.

Auscher's case is probably the most instructive one on record, for the sclerosis was practically limited to the posterior columns, while the symptoms of Friedreich's ataxia were well shown. The patient had inco-ordination, but sensibility was normal. In other words, the various forms of sensation were transmitted otherwise than by the posterior columns. When we consider the other autopsies the same thing is true, with the added fact of the almost invariable sclerosis of the direct cerebellar tract. In these cases, again, sensation remains practically unimpaired. In short, when all the tracts which degenerate upward are sclerosed, we find that patients are still able to secure conduction of sensory impulses; but in all alike we find the primary, the most important symptom, to be essentially a motor disturbance.

Notice also that the later and accessory symptoms are also motor. The speech defect, the scoliosis, the club-foot, the nystagmus—all are due to motor defect.

That the inco-ordination is not due to the sclerosis of the crossed pyramidal tracts is also demonstrated by Auscher's case.

All autopsies practically agree that the sclerosis of these tracts is less intense than in the posterior and cerebellar columns.

It would seem, from Auscher's case, to be probable that the sclerosis of the pyramidal tracts is a secondary and late process which does not reach the same completeness as does the destructive process in the other white columns.

In applying these facts to the elucidation of the functions of the cord, it has seemed to me that we must revise the current views upon the functions of the posterior columns, and possibly of the direct cerebellar.

Waller's discovery of the degeneration of system tracts has been invaluable as a means of differentiating the various tracts. It has given us a means of demonstration which far excels in accuracy any other method of dissection, but I contend that we have misinterpreted the facts revealed by the study of Wallerian degeneration. The tracts which degenerate upward are the columns of Goll, Burdach and the direct cerebellar. The assumption has been, and is, that such tracts convey impulses upward only.

There is no necessary connection between the course of the degeneration and the conduction of physiological impulses. It is simply a question of fact whether the two processes proceed in similar or opposite directions. That the three tracts named degenerate upward cannot be disputed.

The question is, do they convey any or all forms of sensory impulse upward, or do they convey other impulses downward?

It seems to me that the recorded autopsies of Friedreich's disease considered in connection with the symptomatology of the disease afford a demonstration that they do not convey upward.

We are driven to conclude that the function of the posterior columns and direct cerebellar is not to carry sensory impulses upward; and, certainly, by the facts of Friedreich's ataxia we are equally compelled to believe that they convey motor impulses of some sort. If motor, then in all probability they are downward impulses. That they are the main tracts for the conveyance of co-ordinated motor impulses seems to be the necessary corollary of the facts and symptoms of Friedreich's disease. The facts of embryology certainly tend to strengthen the theory. At the end of foetal life, at a time when the crossed pyramidal tracts are undeveloped, the posterior columns and cerebellar tracts are complete. Their function evidently begins at once after

birth. When we remember that the new born infant is characterized not by voluntary control of its muscles, not by accuracy of sense, perception, but by an extensive co-ordination of involuntary motor functions, the conclusion is easy that these tracts subserve these purposes.

Indeed, there are no other tracts developed at birth by which such co-ordination could be carried on.

To sum up: I would conclude that the posterior and direct cerebellar tracts do not convey any sensory impulses upward, except possibly that of the muscular sense.

2d. That the direction of Wallerian degeneration does not necessarily coincide with the usual direction of physiological impulses.

3d. That the posterior columns are the tracts by which co-ordinating motor impulses are sent downward from the cerebellum and mid-brain, and from higher to lower levels of the cord.

4th. That this theory of the function of the posterior and cerebellar columns is the only one which coincides with the embryological development, and is further borne out by the anatomical relations of the superior terminations of these tracts. Their connection with the cerebellum and basal ganglia is direct, with the sensory tracts in the internal capsule extremely uncertain.